Case Report
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Polypoid Ganglioneuroma in a Patient with Colonic Polyposis: A Case Report

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Abstract

Ganglioneuromas are benign tumors of the autonomic nervous system. They are rarely found in the colonic mucosa, where symptoms tend to be non-specific. Some patients may present with abdominal pain, changes in bowel habits, hematochezia, ileus, or obstruction. There are few reports in the literature of ganglioneuromas coexisting with colonic polyposis or even adenocarcinoma. We report a case of a 37-year-old man with polypoid ganglioneuroma, colonic polyposis, and a family history of colon cancer, who had a favorable outcome after two years of follow-up. Ganglioneuroma is a neuroectodermal tumor that is rarely observed in the colorectal mucosa. In the case of polypoid ganglioneuroma, as seen in our patient, no syndromic associations were identified. The patient was successfully treated with endoscopic polypectomy, resulting in a favorable clinical outcome.

Keywords: Ganglioneuroma, Ascending colon, Colonoscopy, Polyposis, Case report

Introduction

Ganglioneuromas are slow-growing hamartomatous tumors that are infrequently found in the colonic mucosa.[1] Three types of these tumors are recognized in the gastrointestinal tract: polypoid ganglioneuroma, ganglioneuromatous polyposis, and diffuse ganglioneuromatosis.[2] These types are differentiated based on specific endoscopic and pathological characteristics. Patients with colonic ganglioneuromas are typically asymptomatic; however, they may present with nonspecific symptoms such as abdominal pain, irritable bowel syndrome, hematochezia, or megacolon with intestinal obstruction.[3] The few cases reported in medical literature have generally been benign; however, some reports indicate associations with colonic polyposis and colorectal cancers.[4] We present a case of polypoid ganglioneuroma in a patient with colonic polyposis who underwent surveillance colonoscopy.

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Case Presentation

A 37-year-old man presented with hematochezia and iron deficiency anemia. His past medical history was unremarkable except for hypothyroidism diagnosed 10 years prior, and he denied any history of smoking or substance abuse. Physical examination revealed only mild pallor. His family history included colorectal cancer in both his aunt and grandmother. Laboratory tests indicated mild microcytic hypochromic anemia, hypothyroidism, and a positive fecal occult blood test. Abdominopelvic ultrasonography revealed a 24 × 20 mm hyperechoic lesion, suggesting a hemangioma in the left lobe of the liver.

He was referred for upper and lower gastrointestinal tract endoscopy. Upper endoscopy revealed multiple small polyps in the distal part

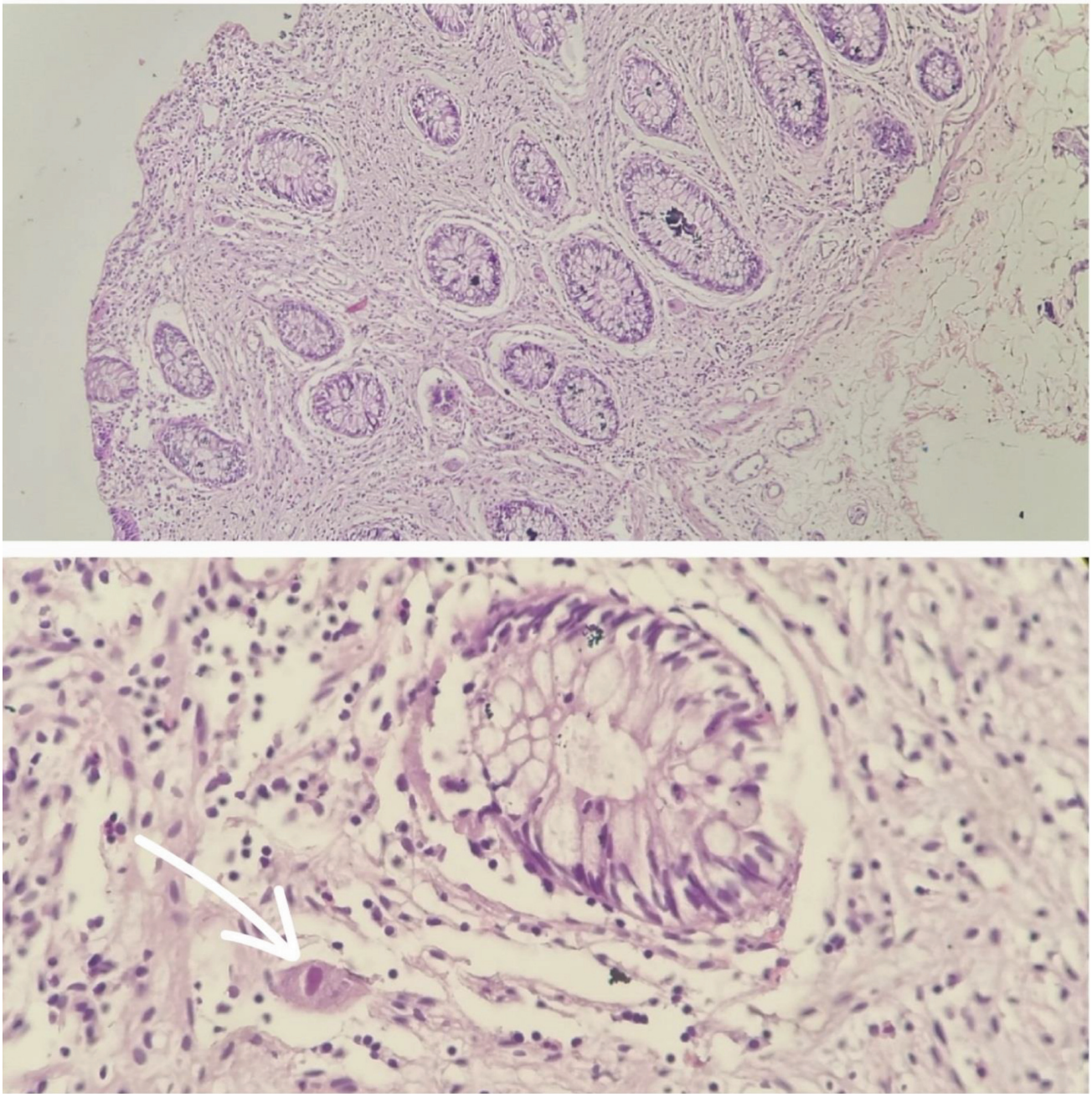

Figure 1. Hematoxylin and eosin (H&E) microscopic image: The image illustrates the proliferation of spindle-shaped cells with wavy nuclei (Schwann cells) and scattered large cells with eccentric vesicular nuclei, prominent nucleoli, and abundant amphophilic cytoplasm (ganglion cells, arrow) (Upper, 100× Lower, 400× magnification).

of the body, antrum, and prepyloric areas, which were confirmed to be hyperplastic polyps upon histopathological examination. Colonoscopy showed one large pedunculated polyp in the ascending colon and multiple sessile polyps of varying diameters throughout the colon. These were identified as adenomatous polyps (tubulovillous type with low-grade dysplasia) and multiple sessile inflammatory polyps on histological examination.

The patient was followed up with regular surveillance colonoscopies at one-year intervals. A surveillance colonoscopy performed one year later revealed numerous small polyps in the rectum and other parts of the colon, as well as one 1 × 1 cm sessile polyp in the ascending colon. This latter polyp was removed using a hot snare after methylene blue injection. Histopathological examination showed pieces of colonic mucosa where the lamina propria was expanded due to a proliferation of spindled cells accompanied by aggregates of large cells resembling ganglion cells (Figure 1). Immunohistochemical studies confirmed the diagnosis of ganglioneuroma, with S100 marker staining the Schwann and ganglion cells (Figure 2). Over the next two years, the patient reported no complaints; however, surveillance colonoscopies revealed adenomatous polyps with low-grade dysplasia in the transverse colon and rectum.

Ethics approval

This study was approved by the Institutional Review Board of the Shiraz University of Medical Science (Approval ID: IR.SUMS.MED.REC. 1403.602).

Discussion

Ganglioneuromas are benign, slow-growing tumors of neuroectodermal origin and are classified within a group of tumors known as peripheral neuroblastic tumors, which also includes neuroblastomas, ganglioneuroblastomas, and ganglioneuroma.[5] They typically arise in the adrenal gland and retroperitoneal ganglia but can also be found in the head and neck or posterior mediastinum, where major sympathetic ganglia are located.[3,6] They are rare in the gastrointestinal tract, where they may present with symptoms such as abdominal pain, changes in bowel habits, weight loss, hematochezia, ileus, or even obstruction.[1,6] The occurrence and severity of these symptoms depend on the size, location, and type of polyps involved. Iwamuro et al. reported a case of a patient with neurofibromatosis type 1 and diffuse ganglioneuromatosis who presented with multiple colorectal ulcers, as well as concomitant anal fistula and a history of intra-abdominal abscess.[7]

Three types of ganglioneuromatous diseases are described in the gastrointestinal tract: polypoid ganglioneuroma, ganglioneuromatous polyposis, and diffuse ganglioneuromatosis. All of these conditions predominantly affect the colon and rectum.[2] The first and most common type is characterized by small (usually less than 2 cm), single or multiple polyps located in the mucosa and submucosa of the colon. These polyps may have either sessile or pedunculated morphology and cannot be differentiated from adenomatous or hyperplastic polyps based solely on endoscopic examination. Histologically, they resemble juvenile polyps, exhibiting disturbed crypt architecture and cystically dilated glands, while the lamina propria is expanded by a proliferation of ganglion cells interspersed with nerve fibers and Schwann cells.[4] The second type has similar histology but presents with numerous (typically more than 20) polyps of equal size (usually less than 2 cm). The third type, diffuse ganglioneuromatosis, is characterized by large (ranging from 1 to 17 cm in diameter), nodular, mucosal or transmural lesions that are ill-defined and can involve the myenteric plexus.[4,5]

Ganglioneuromas may occur sporadically or in association with genetic syndromes. Single lesions such as polypoid ganglioneuromas are not typically associated with genetic syndromes and generally have a favorable prognosis, with a low tendency to recur.[1,4] In contrast, ganglioneuromatous polyposis has been linked to familial adenomatous polyposis (FAP), Cowden's disease, tuberous sclerosis, multiple endocrine neoplasia type 2b (MEN IIb) syndrome, colorectal carcinoma, and juvenile polyposis. Diffuse gan-

glioneuromatosis usually occurs in the context of neurofibromatosis type 1 (also known as von Recklinghausen disease) or MEN IIb (which includes parathyroid adenoma and medullary thyroid carcinoma).[4]

There have been a few cases where colon cancer coexists with ganglioneuromatous polyposis or diffuse ganglioneuromatosis in the same patient; however, no established association exists between gastrointestinal ganglioneuromas and malignant tumors.[3] Oh et al. suggested that diffuse ganglioneuromatosis could be a premalignant condition based on their report of a 26-year-old male patient who developed colon cancer 12 years after being diagnosed with colonic diffuse ganglioneuromatosis.[3] They proposed that persistent chronic inflammation in such patients might trigger colorectal cancer, similar to colitis-related cancer caused by chronic inflammatory disease. Given the high risk of cancer associated

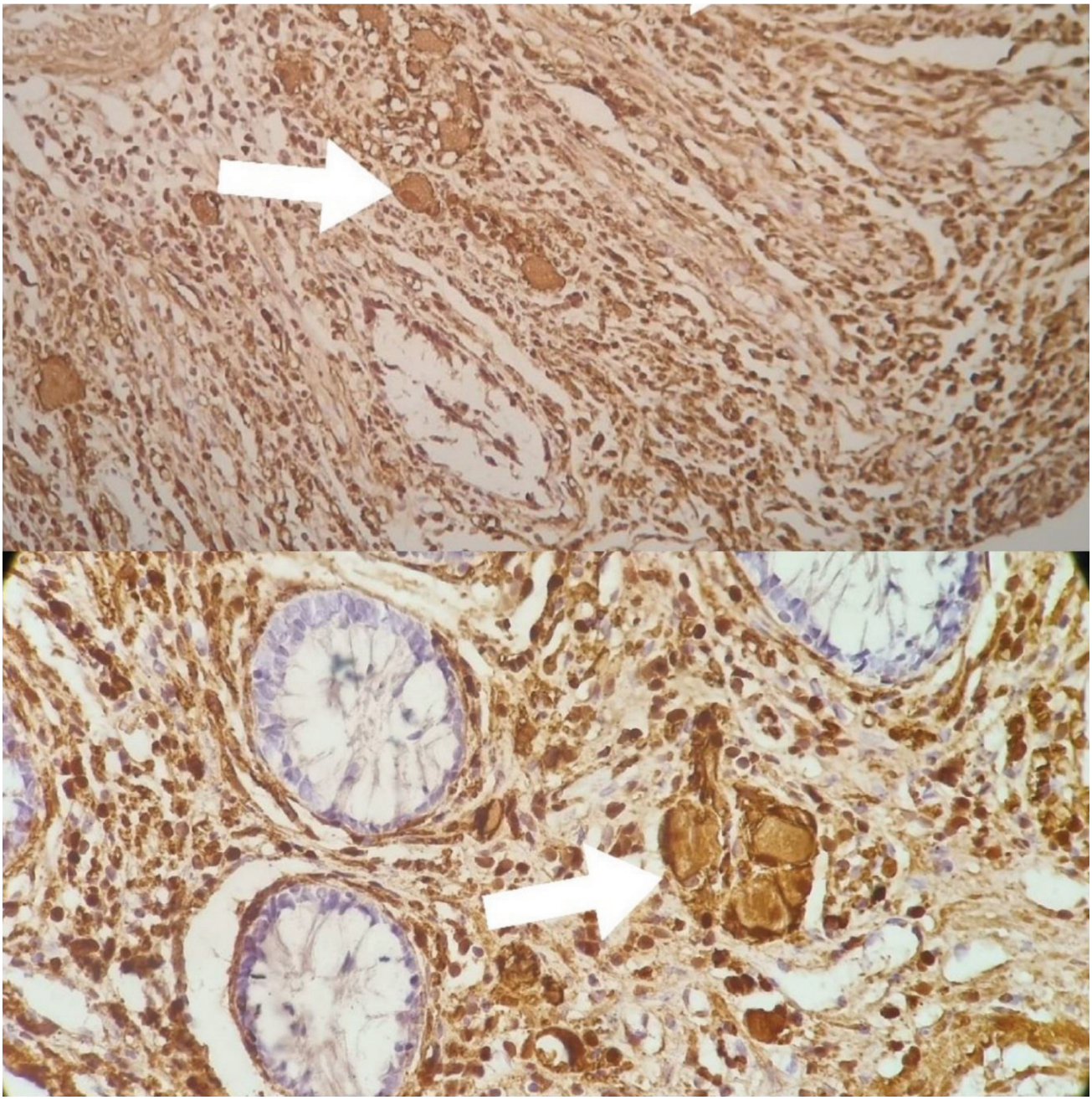

Figure 2. S100 protein immunohistochemistry (IHC) stain showing cytoplasmic and nuclear immunoreactivity in ganglion cells (arrows) and Schwann cells (100× magnification).

with syndromes linked to ganglioneuromas, patients should be screened for tumors in the colon, thyroid, breast, and uterus.[6]

Colonic ganglioneuromas do not have distinct endoscopic features, and a definitive diagnosis can only be made through histopathological examination.1 Generally, hematoxylin and eosin-stained sections are sufficient for histological diagnosis.[3] However, immunohistochemical studies can be helpful in confirming the neural origin of these lesions using markers such as neurofilaments, synaptophysin, chromogranin, S100 protein (Figure 2), glial fibrillary acidic protein (GFAP), CD56, and neuron-specific enolase.[5]

Solitary ganglioneuromas have a favorable prognosis, and patients are not predisposed to developing multiple endocrine neoplasia.[7] Given their association with various syndromes, genetic counseling is recommended based on additional signs and symptoms of the patient and their family history. Endoscopic resection of the polyp is the only treatment required.[3]

There is no evidence regarding the effectiveness of regular surveillance colonoscopy, and a definitive treatment approach has not been well established. Most clinicians agree that repeated colonoscopy is unnecessary following endoscopic resection.[8] However, Oh et al. recommend a surveillance colonoscopy interval of 1 to 2 years.3 Many studies, including one by Arai et al., suggest that endoscopic mucosal resection is an effective therapy.[1]

Conclusion

Ganglioneuroma is a neuroectodermal tumor that is rarely found in the colorectal mucosa. Due to its rarity, the exact incidence, presentation, and natural course are not well understood. There are currently no established guidelines for monitoring and managing these patients. When a gastrointestinal polyp is identified as a ganglioneuroma, the patient should be evaluated for genetic syndromes and associated cancers, despite the benign nature of these tumors. In cases of polypoid ganglioneuroma, such as ours, no syndromic association was found. Our patient was successfully treated with endoscopic polypectomy and had a favorable clinical outcome. Further studies are needed to establish guidelines for the management of these tumors and treatment recommendations.

Informed Consent

Written informed consent was obtained from the patient's legal guardian for publishing this case report and any accompanying images. A copy of the written consent is available upon the request of the Editor-in-Chief of the journal.

Availability of Data and Material

The data details were presented in the case presentation section.

Authors' Contributions

F.J, F.K, and F.A and M.K: Study design; data acquisition; data analysis and interpretation; drafting and critical reviewing of the manuscript. All authors read and approved the final manuscript version and agreed with all parts of the work in ensuring that any queries about the accuracy or integrity of any component of the work are appropriately investigated and handled.

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Funding

None declared.

Conflict of Interest

None declared.

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